RX.PA.054.CCH SOLIRIS (ECULIZUMAB) & ULTOMIRIS (RAVULIZUMAB-CWVZ)

The purpose of this policy is to define the prior authorization process for Ultomiris® (ravulizumab-cwvz) and Soliris® (Eculizumab)

DEFINITIONS

ADAMTS13 – A Disintegrin And Metalloprotease with a ThromboSpondin type 1 motif, member 13. A deficiency in ADAMTS13 protease can lead to Thrombotic Thrombocytopenic Purpura (TTP).

Myasthenia Gravis Foundation of America (MGFA) Disease Clinical Classification

Class I	Any ocular muscle weakness; may have weakness of eye closure. All other muscle strength is normal.
Class II	Mild weakness affecting muscles other than ocular muscles; may also have ocular muscle weakness of any severity. • IIa. Predominantly affecting limb, axial muscles, or both. May also have lesser involvement of oropharyngeal muscles. • IIb. Predominantly affecting oropharyngeal, respiratory muscles, or both. May also have lesser or equal involvement of limb, axial muscles, or both.
Class III	Moderate weakness affecting muscles other than ocular muscles; may also have ocular muscle weakness of any severity. • IIIa. Predominantly affecting limb, axial muscles, or both. May also have lesser involvement of oropharyngeal muscles. • IIIb. Predominantly affecting oropharyngeal, respiratory muscles, or both. May also have lesser or equal involvement of limb, axial muscles, or both.
Class IV	Severe weakness affecting muscles other than ocular muscles; may also have ocular muscle weakness of any severity. • IVa. Predominantly affecting limb, axial muscles, or both. May also have lesser involvement of oropharyngeal muscles. • IVb. Predominantly affecting oropharyngeal, respiratory muscles, or both. May also have lesser or equal involvement of limb, axial muscles, or both
Class V	Defined as intubation, with or without mechanical ventilation, except when employed during routine postoperative management. The use of a feeding tube without intubation places the patient in class IVb.

POLICY

It is the policy of the Health Plan to maintain a prior authorization process that promotes appropriate utilization of specific drugs with potential for misuse or limited indications. This process involves a review using Food and Drug Administration (FDA) criteria to

make a determination of Medical Necessity, and approval by the Medical Policy Committee.

The drugs, Ultomiris® (ravulizumab-cwvz) and Soliris® (Eculizumab), is subject to the prior authorization process.

PROCEDURE

I. PLAN DESIGN SUMMARY

Requests for Ultomiris and Soliris are subject to the preferred medical drug list program. This program applies to the products specified in this policy. Coverage for non-preferred products is provided based on clinical circumstances that would exclude the use of the preferred product and may be based on previous use of a product. The coverage review process will ascertain situations where a clinical exception can be made. Each referral is reviewed based on all utilization management (UM) programs implemented for the client

	Product
Preferred	Ultomiris® (ravulizumab-cwvz)
Non-preferred	Soliris® (Eculizumab)

II. EXCEPTION CRITERIA (Use for non-preferred request only)

This program applies to members requesting treatment for an indication that is FDA-approved for the preferred product(s) (as applicable).

Coverage for the non-preferred product is provided when ANY of the following criteria are met:

- Member has an inadequate response with the preferred product
- Member has a documented intolerable adverse event with the preferred product
- Member has a contraindication to the preferred product

III. CLINICAL CRITERIA BELOW (Use for ALL drug request)

Initial Authorization Criteria:

Must meet criteria for all diagnosis in addition to that listed under the respective diagnosis:

For All Diagnosis:

- Prescriber must be enrolled in Ultomiris and Soliris Risk Evaluation and Mitigation Strategies (REMS)
- Must be prescribed at a dose within the manufacturer's dosing guidelines (based on diagnosis, weight, etc.) listed in the FDA approved labeling
- Must be prescribed by or in consultation with a hematologist, oncologist, neurologist, immunologist, or genetic specialist
- Must not be used in combination with other biologic therapies used for the given diagnosis

1. Paroxysmal Nocturnal Hemoglobinuria (PNH)

- Must be the following age, depending on drug requested:
  - At least 1 month of age or older (Ultomiris)
  - At least 18 years of age or older (Soliris)
- Must have a laboratory confirmed diagnosis of paroxysmal nocturnal hemoglobinuria (PNH) as evidenced by having detectable GPI-deficient hematopoietic clones (Type III PNH RBC) via Flow Cytometry. Documentation of Flow Cytometry pathology report support must indicate presence of PNH-type RBC (red blood cell) and must be submitted.
- Must have an LDH level 1.5 times the upper limit of the normal range (laboratory result with reference range must be submitted) and at least ONE sign or symptom of disease, such as:
  - Thromboembolic events
  - Symptomatic anemia
  - Organ damage secondary to chronic hemolysis (e.g., renal insufficiency, pulmonary insufficiency)
  - Abdominal pain
  - Fatigue
  - Erectile Dysfunction
  - *EXCEPTION:* Members who are pregnant do not require a sign/symptom so long as provider deems potential benefit outweighs potential fetal risk
- Must have documentation of the following baseline laboratory values or information:
  - Hemoglobin level
  - Current RBC transfusion requirements
  - History of thrombotic events
- **For non-preferred products:** Must have a trial and failure of the preferred product

2. Myasthenia gravis (MG)

- Must have a diagnosis of generalized myasthenia gravis, with documentation of a positive test for anti-acetylcholine receptor (AChR) antibody

- Must be age 18 years or older
- Must have a Myasthenia Gravis Foundation of America (MGFA) Clinical Classification of class II to IV
- Must have a Myasthenia Gravis-Specific Activities of Daily Living (MG-ADL) total score of ≥ 6
- Must rhave a trial and failure, contraindication, or intolerance to at least ONE corticosteroid (e.g., prednisone)
- Must have a trial and failure or contraindication to at least ONE of the following (or intolerance of all):
  - At least TWO non-steroidal immunosuppressive therapies (e.g., azathioprine, cyclophosphamide, methotrexate)
  - At least ONE non-steroidal immunosuppressive therapy while on chronic plasmapheresis or plasma exchange
- **For non-preferred products:** Must have a trial and failure of the preferred product

3. Neuromyelitis Optica Spectrum Disorder (NMOSD)

- Must have a diagnosis of neuromyelitis optica spectrum disorder (NMOSD)
- Must have documentation of a positive blood serum test for anti-aquaporin-4 antibodies (AQP4-IgG)
- Must be age 18 years or older
- Must have a history of ONE of the following:
  - At least 2 relapses in last 12 months
  - At least 3 relapses in the last 24 months, with at least 1 relapse in the 12 months prior to request
- Must have a baseline Expanded Disability Status Scale (EDSS) score of ≤ 7 (consistent with the presence of at least limited ambulation with aid)
- **For non-preferred products:** Must have a trial and failure of the preferred product

4. Atypical hemolytic uremic syndrome (aHUS)

- Must be the following age, depending on drug requested:
  - At least 1 month of age or older (*Ultomiris)*
  - At least 2 months of age or older (*Soliris)*
- Must have documentation or attestation from the provider of the following:
  - Thrombotic Thrombocytopenic Purpura (TTP) has been ruled out by evaluating ADAMTS-13 level (i.e., ADAMTS-13 activity level ≥ 10%)
  - Shiga toxin E. coli related to hemolytic uremic syndrome (STEC-HUS) has been ruled out
- Must provide documentation baseline serum LDH, platelet count, serum creatinine, and plasma exchange requirements

- **For non-preferred products:** Must have a trial and failure of the preferred product

Reauthorization Criteria:

All prior authorization renewals are reviewed on an annual basis to determine the Medical Necessity for continuation of therapy. Authorization may be extended at 1-year intervals based upon chart documentation of the following:

- **Paroxysmal Nocturnal Hemoglobinuria (PHN)**
  - Documentation from the prescriber that member's condition has improved, as evidenced by one of the following: decrease in serum LDH, Hgb level above baseline, or reduction in need for blood transfusions
  - Must be prescribed at a dose within the manufacturer's dosing guidelines (based on diagnosis, weight, etc.) listed in the FDA approved labeling
  - Must not be used in combination with other biologic therapies used for PHN

- **Myasthenia Gravis**
  - Reduction in MG-ADL assessment score from pretreatment baseline
    - For subsequent reauthorizations, stability in the score may be accepted after initial improvement
  - Must be prescribed at a dose within the manufacturer's dosing guidelines (based on diagnosis, weight, etc.) listed in the FDA approved labeling
  - Must not be used in combination with other biologic therapies used for myasthenia gravis

- **Neuromyelitis Optica Spectrum Disorder (NMOSD)**
  - Documentation from prescriber that member's condition has improved as evidenced by at least ONE of the following:
    - Decrease in frequency of relapse
    - Reduction in EDSS score from baseline
    - Reduced hospitalizations
  - Must be prescribed at a dose within the manufacturer's dosing guidelines (based on diagnosis, weight, etc.) listed in the FDA approved labeling
  - Must not be used in combination with other biologic therapies used for NMOSD

- **Atypical Hemolytic Uremic Syndrome (aHUS)**
  - Documentation from prescriber showing that the member's condition has improved, as evidence by at least ONE of the following:
    - Decrease in serum LDH

        - Improvement in serum creatinine
        - Normalization of platelet counts
        - Decrease in plasma exchange
    - Must be prescribed at a dose within the manufacturer's dosing guidelines (based on diagnosis, weight, etc.) listed in the FDA approved labeling
    - Must not be used in combination with other biologic therapies used for aHUS

Limitations:

Length of Authorization (if above criteria met)	
Initial Authorization	Up to 1 year
Reauthorization	Same as initial

If the established criteria are not met, the request is referred to a Medical Director for review, if required for the plan and level of request.

Codes: J Code(s)

Code	Brand	Description
J1300	Soliris	INJECTION, ECULIZUMAB, 10MG
J1303	Ultomiris	INJECTION RAVULIZUMAB-CWVZ 10 MG

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REVIEW HISTORY

DESCRIPTION OF REVIEW / REVISION	DATE APPROVED
Initial Review	3/22
Updated initial authorization duration to 1 year	02/23
Addition of Soliris Addition of 2 indications: NMOSD & MG	08/2024

Record Retention

Records Retention for Evolent Health documents, regardless of medium, are provided within the Evolent Health records retention policy and as indicated in CORP.028.E Records Retention Policy and Procedure.

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